# PART XIV Immunology

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1. A 9-month-old boy presented with recurrent attacks of diarrhea, thrush, and failure to thrive shortly after birth. You suspect immune deficiency.
Of the following, the **MOST** common cause of this condition is

A. hyper-IgE syndrome
B. defect in phagocytic cells
C. deficiencies in T-cell function
D. defect in antibody production
E. defect in complement proteins

2. A 17-month-old girl presented with history of recurrent attacks of pneumonia since the age of 8 months. You suspect a predominant B-cell defect.
Of the following, the **BEST** simple initial screening test is

A. IgA measurement
B. IgG measurement
C. IgM measurement
D. IgE measurement
E. isohemagglutinins titer

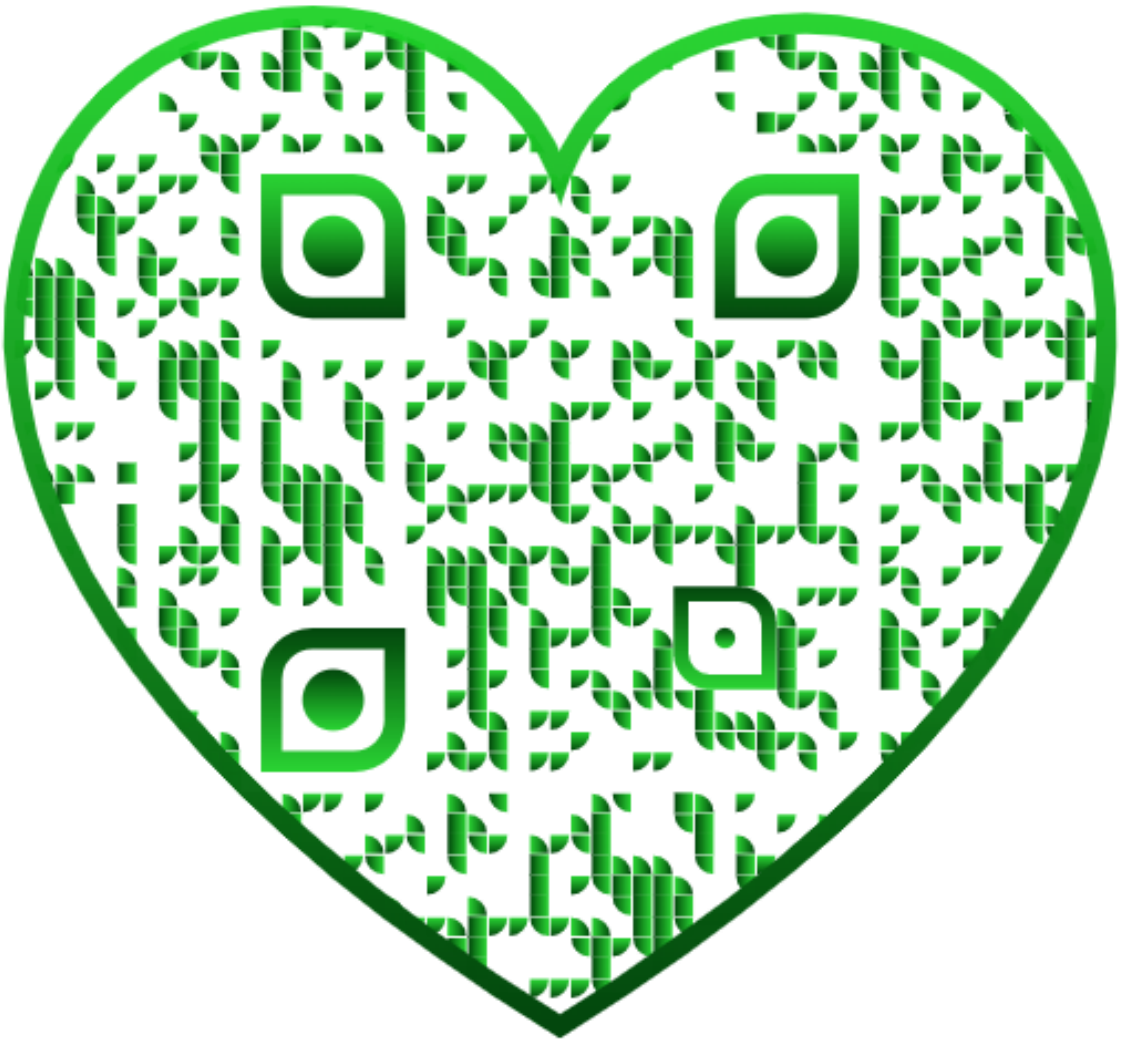
**selective IgA deficiency, which is the most common Bcell defect, is excluded, as are most of the permanent types of hypogammaglobulinemia, as IgA is usually very low or absent in those conditions. If IgA is low, IgG and IgM should also be measured.**

3. A very high serum concentrations of one or more immunoglobulin classes, suggest all the following **EXCEPT**

A. HIV infection
B. chronic inflammation
C. intestinal lymphangiectasia
D. chronic granulomatous disease
E. autoimmune lymphoproliferative syndrome

4. A 3-year-old boy presented with recurrent attacks of pneumonia and otitis media since the 1st birthday. Your diagnosis is X-linked agammaglobulinemia.
Of the following, the **MOST** likely offending organism is

A. CMV
B. Mycoplasma
C. Pneumocystis jiroveci
D. Staphylococcus aureus
E. Streptococcus pneumonia



5. A 4.5-year-old boy presented with history of recurrent attack of secretary otitis media and purulent nasal discharge, Haemophilus influenza revealed by culture of ear discharge, on physical examination there is no tonsillar tissue and no palpable lymph nodes.
Of the following, the **MOST** appropriate test to confirm the diagnosis is measurement of

A. flow cytometry
B. IgA concenteration
C. isohemagglutinins titer
D. IgG and IgM concenteration
E. antibodies to antigens of routine immunizations

**The diagnosis of X-linked agammaglobulinemia (XLA) should be suspected if lymphoid hypoplasia is found on physical examination (minimal or no tonsillar tissue and no palpable lymph nodes). Flow cytometry is an important test to demonstrate the absence of circulating B cells, which will distinguish this disorder from common variable immunodeficiency, the hyper-IgM syndrome, and transient hypogammaglobulinemia of infancy.**

6. A 3-year-old girl presented with recurrent attacks of chest infection and frequent episodes of chronic diarrhea, stool examination revealed Giardia lamblia and serum IgA level was very low.
Regarding this condition, all the following are true **EXCEPT**

A. autosomal dominant inheritance
B. phenotypically normal blood B cells
C. administration of IVIG is not indicated
D. environmental factors may trigger the disease
E. it is associated with a celiac-like syndrome with dramatic response to gluten-free diet

**IgA deficiency is associated with a celiac-like syndrome, which may or may not respond to a gluten-free diet.**

7. A 2-year-old boy presented with failure to thrive, neonatal hypocalcemic seizure, and three attacks of sepsis. Examination reveals cleft palate and holo-systolic murmur.
Of the following, the genetic predisposition of this condition is

A. microdeletions of chromosome 22q11.2
B. interstitial deletion of the long arm of chromosome 15 (15q11-13)
C. presence of two copies of paternally derived chromosome 11p15.5
D. presence of mutated gene on long arm of chromosome 11 (11q22-23)
E. presence of abnormal gene, on the proximal arm of the X chromosome at Xp11.22-11.23

**A. DiGeorge syndrome,
B. Prader-Willi syndrome,
C. Beckwith-Wiedemann syndrome,
D. Wiskott-Aldrich syndrome,
E. ataxia-telangiectasia.**

8. Of the following, the findings that are **MOST** suggestive of DiGeorge syndrome are

A. cataract and a congenital heart defect
B. hypotonia and a congenital heart defect
C. cutis aplasia and a congenital heart defect
D. hypocalcemia and a congenital heart defect
E. mongoloid slant to the eyes and a congenital heart defect

9. The **MOST** appropriate method to correct the immune deficiency in complete DiGeorge syndrome is

A. vaccination
B. administration of IVIG
C. judicious use of antibiotics
D. transplantation of thymic tissue
E. transplantation of hematopoietic stem cells

10. In addition to infection and bleeding, the **MOST** common cause of death in Wiskott-Aldrich syndrome is
A. renal failure
B. graft versus host disease
C. protracted bloody diarrhea
D. EBV-associated malignancies
E. development of autoimmune disease

11. The treatment of choice for a 3-year-old boy with Wiskott-Aldrich syndrome is
A. splenectomy
B. use of killed vaccines
C. aggressive antibiotics
D. administration of IVIG
E. bone marrow or cord blood transplantation

12. All the following are features of ataxia-telangiectasia **EXCEPT**
A. hepatic manifestation
B. high incidence of malignancy
C. chronic sinopulmonary disease
D. oculocutaneous telangiectasias
E. variable humoral and cellular immunodeficiency

13. The earliest clinical feature of ataxia-telangiectasia is
A. ataxia
B. jaundice
C. inability to walk
D. ocular telangiectasias
E. cutaneous telangiectasias

14. Severe eosinophilia, defined as an absolute eosinophil count (AEC) more than
A. 750 cells/µL
B. 1,000 cells/µL
C. 1,500 cells/µL
D. 5,000 cells/µL
E. 7,500 cells/µL

15. Which of the following statements is **TRUE** regarding diurnal variation of the absolute eosinophil count (AEC)?

A. higher in the evening
B. lower in the afternoon
C. higher in the afternoon
D. lower in the early morning
E. higher in the early morning

16. Eosinophilia can be seen in all the following **EXCEPT**

A. scabies
B. urticaria
C. corticosteroid therapy
D. Hodgkin disease
E. hypersensitivity drug reactions

17. The following conditions can cause eosinophilia **EXCEPT**

A. malaria
B. filariasis
C. giardiasis
D. ascariasis
E. amebiasis

**Eosinophilia are caused by tissue-invasive as helminth infections; ascariasis, filariasis, amebiasis, malaria, and scabies whereas entrobius vermicularis and Giardia lamblia are solely intraluminal in gastrointestinal tract.**

18. The highest white blood cells count is seen at

A. birth
B. 12 hour of life
C. 1 week of life
D. 1 year of life
E. adolescence

19. Severe neutropenia is defined as absolute neutrophil count (ANC) less than

A. 500/µL
B. 750/µL
C. 1000/µL
D. 1250/µL
E. 1500/µL

20. Chronic neutropenia by definition lasts longer than

A. 1 mo
B. 3 mo
C. 6 mo
D. 9 mo
E. 12 mo

21. Chronic neutropenia may be induced by
    A. adenovirus
    B. enteroviruses
    C. cytomegalovirus
    D. influenzas A and B
    E. respiratory syncytial virus

22. A 4-year-old boy presented with high fever, decreased appetite, aphthous stomatitis, and cellulitis of the right calf muscle. His absolute neutrophil count (ANC) was 600/µL.
The **MOST** likely organisms causing this condition are
    A. Staphylococcus aureus and Histoplsmosis
    B. Staphylococcus aureus and rickettsial pox
    C. Staphylococcus aureus and Cytomegalovirus
    D. Staphylococcus aureus and Pneumocystis carinii
    E. Staphylococcus aureus and gram-negative bacteria

23. Recurrent infections with neutropenia are a distinctive feature of
    A. Pompe disease
    B. Hurler disease
    C. Gaucher disease
    D. Glycogen storage disease Ia (GSDIa)
    E. Glycogen storage disease Ib (GSDIb)

24. All the following drugs can induce neutropenia **EXCEPT**
    A. penicillin
    B. tetracycline
    C. phenobarbital
    D. phenothiazine
    E. chloramphenicol

25. A 15-year-old boy with rheumatoid arthritis, on quinine for 6 months, presented with fever, aphthous stomatitis, and fatigue for the last 5 days. His absolute neutrophil count (ANC) was 700/µL.
The **MOST** next appropriate therapeutic measure to be done when quinine withdrawal fails to improve neutropenia is
    A. administration of IVIG
    B. aggressive use of antibiotics
    C. transplantation of hematopoietic stem cells
    D. administration of pulse methylprednisolone
    E. administration of recombinant human granulocyte colony-stimulating factor

26. A 9-year-old boy with acute lymphoblastic leukemia on chemotherapy, presented with fever, aphthous stomatitis, and fatigue for the last 5 days. His absolute neutrophil count (ANC) was 450/µL and hemoglobin level was 10gm/dl.
The **NEXT** appropriate therapeutic measure is

A. administration of IVIG
B. transplantation of hematopoietic stem cells
C. administration of pulse methylprednisolone
D. aggressive use of broad-spectrum antibiotics
E. administration of recombinant human granulocyte colony-stimulating factor

27. A 13-month-old girl presented with failure to thrive, repeated chest infection, and fatty offensive diarrhea. Hemoglobin level 8.8gm/dl, platelate count 150/mm3, white blood cell count 3.500/mm3, with absolute neutrophil count (ANC) 500/µL.
Of the following, the **MOST** likely diagnosis is

A. Kostmann disease
B. cyclic neutropenia
C. drug-induced neutropenia
D. acute lymphoblastic leukemia
E. Shwachman-Diamond syndrome

28. Leukemoid reactions (WBC count > 50,000/µL) are characterized by abundant

A. blasts
B. band form
C. myelocytes
D. promyelocytes
E. metamyelocytes

29. Leukemoid reactions (WBC count >50,000/µL) are caused by all the following **EXCEPT**

A. shigellosis
B. septicemia
C. salmonellosis
D. chronic granulomatous disease
E. leukocyte adhesion defect with infection

30. Shifting to the left (proportion of immature neutrophil cells >5%) may occasionally be caused by all the following **EXCEPT**

A. burns
B. trauma
C. hemorrhage
D. acute hemolysis
E. megaloblastic anemia **Hypersegmented neutrophils**

31. A 15-year-old boy presented with high fever, lymphadenopathy, hepatosplenomegaly, recurrent skin infections, silvery hair, and photophobia. Last year, he developed ataxia.
The **MOST** characteristic finding in this condition is

A. neutropenia
B. thrombocytopenia
C. prolonged bleeding time
D. prolonged prothrombine time
E. prolonged partial prothrombine time

**Patients with Chédiak-Higashi syndrome (CHS) have prolonged bleeding times with normal platelet counts, resulting from impaired platelet aggregation associated with a deficiency of the dense granules containing adenosine diphosphate and serotonin.**

32. In patient with Chédiak-Higashi syndrome, the only curative therapy is hematopoietic stem cell transplantation which correct all the following **EXCEPT**

A. neuropathy
B. immunologic function
C. hematopoietic function
D. natural killer cell deficiency
E. conversion to the accelerated phase

33. A 3-year-old boy presented with recurrent subcutaneous abscesses, lymphadenitis, osteomyelitis, and family history of recurrent infections.
The hallmark of this condition is

A. hypocalcemia
B. atopic dermatitis
C. thrombocytopenia
D. granuloma formation
E. prolonged bleeding time

34. The **MOST** common pathogen in chronic granulomatous disease (CGD) is

A. S. aureus
B. Aspergillus
C. Salmonella
D. Mycobacterium
E. Candida albicans

35. The diagnosis of chronic granulomatous disease CGD is **MOST** often made by

A. DNA analysis
B. flow cytometry
C. neutrophil G6PD assay
D. erythrocyte G6PD assay
E. nitroblue tetrazolium dye test

36. To reduce the number of bacterial infection in patients with chronic granulomatous disease (CGD), they should be given daily

A. penicillin
B. cloxacillin
C. interferon-γ
D. itraconazole
E. trimethoprim-sulfamethoxazole

37. In chronic granulomatous disease (CGD), the best method to detect and follow up deep-seated infection is

A. ESR
B. blood culture
C. WBC count
D. C-reactive protein
E. immature to mature neutrophil ratio

38. All the following are characteristic in properdin deficiency **EXCEPT**

A. dermal vasculitis
B. male predominance
C. systemic lupus erythematosus
D. susceptibility to N. meningitidis meningitis
E. normal serum hemolytic complement activity

39. All the following matching about genetic deficiencies of plasma complement components and associated clinical findings are true **EXCEPT**

A. C4 ---------- discoid lupus erythematosus
B. C1q---------- systemic lupus erythematosus
C. C1 INH ---------- hereditary angioedema
D. factor H---- atypical hemolytic-uremic syndrome
E. properdin ---------- N. meningitidis meningitis

40. The complement component of both the classical and alternative pathways is

A. C2
B. C3
C. C4
D. C5
E. C6

41. All the following conditions can induce lymphopenia **EXCEPT**

A. sepsis
B. typhoid
C. brucellosis

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**Chronic bacterial infections such as tuberculosis and brucellosis may lead to a sustained lymphocytosis.**

D. corticosteroid use
E. cartilage-hair hypoplasia

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